Novel Association of Elastofibroma with Aortic Stenosis

Report of a Case Report Interfering with a Thoracotomy Procedure and a Reassessment of Typical Patient Demographics and Tumor Location

Barbara Robinson, MD, MS, Ivayla Geneva, MD, PhD,† Steve Landas, MD,‡ Robert Michiel, MD,§ Karen White, MD,‡ Rolfe A. Grage, MD,‖ and Charles Lutz, MD,¶ and William D. Edwards, MD¶*

Elastofibromas are benign connective tissue tumors occurring in elderly patients and usually found in isolation. We report for the first time a case of multiple elastofibromas discovered during thoracotomy in a patient with prior aortic valve replacement for aortic stenosis. We discuss a novel association between aortic stenosis and elastofibroma and stipulate on possible etiology. In light of the well-known association of aortic valve insufficiency and a deficiency in structural proteins, the opposite problem of excess structural proteins may be pathogenic in the formation of both aortic valve stenosis and elastofibromas. This previously unrecognized association may influence the clinical management and diagnostic work-up in patients manifesting with elastofibroma.

Elastofibromas are rare, noncapsulated, slow-growing connective tissue tumors first described by Jarvi and Saxen in 1961,[1] generally benign, but may coexist with sarcomas, found more commonly in middle-aged women, most commonly located in the right infrascapular region, usually asymptomatic, or present with shoulder stiffness, pain, or swelling; generally unilateral although may be bilateral and multiple (Table 1). Small or asymptomatic elastofibromas may be easily overlooked, and on presentation, although most patients feature a several-centimeter-large tumor that does not adhere to the overlying skin and may be deep to chest wall musculature. Elastofibroma is diagnosed clinically with computed tomography (CT) or nuclear magnetic resonance. Immunohistochemically, they express vimentin and produce elastin fibers in characteristic fern-like structures. Because sarcomas may occur in the subscapular site, any suspicious looking lesion on imaging should be confirmed with biopsy to determine extent of chest wall resection necessary. No report exists where elastofibroma, which may have been found accidently during thoracotomy,[2] was associated with aortic stenosis (AS) as in our case. We stipulate on possible common etiology and heightened clinical awareness for associated lesions if elastofibroma is detected.

CASE PRESENTATION

A 76-year-old right-handed white male with cough and history of hypertension, diabetes, alcohol abuse, cholelithiasis, stroke, atrial fibrillation, coronary artery disease, and aortic valve stenosis with valve replacement 2 years ago with #23 mosaic ultra-bioprosthetic valve complained of worsening shortness of breath. He had no prior history of malignancy or trauma. He was previously treated for bronchiolitis with amoxicillin with no resolution. Vital signs were stable. Decreased right-sided breath sounds and a 2×3 cm palpable lateral chest wall mass were noted. Chest radiograph demonstrated a right-sided pleural effusion; he underwent thoracentesis with removal of 700 ml of turbid fluid, but persistent trapped lung, hydropneumothorax, with complex septated loculated pleural collection and empyema on subsequent chest radiograph (Fig. 1*A*) and CT chest (Fig. 1*B*) necessitated prolonged chest tube drainage. tissue plasminogen activator injections failed to dissolve the loculations. During thoracotomy for decortication, a $6.5 \times 5.2 \times 2.2$ cm mobile mass denser than lipoma was palpable within the right lateral chest wall, deep to the latissimus dorsi, requiring resection to access the chest cavity. This tumor was identified on CT scan deep to the right latissimus dorsi muscle (Fig. 1*C*). To gain access to the pleural space for decortication and because of potential for malignancy, the tumor was resected; intraoperative frozen section identified the tumor as a benign elastofibroma and not a sarcoma thus not necessitating further chest wall resection. It measured $6.5 \times 5.2 \times 2.2$ cm (Fig. 2*A*) with nodular fibroadipose and dense, partly cellular collagenous tissue admixed with large, irregular, thick elastic fibers that were deeply eosinophilic on hematoxylin and eosin stain, branching, with areas of globules in a linear arrangement, as "beads on a string"(Fig. 2*B*). An elastic stain highlighted the dense core and irregular margins of the globules (Fig. 2*C*).

Departments of *Surgery-Thoracic Surgery, †Opthalmology, ‡Pathology, §Upstate Heart and Vascular Center, and ‖Radiology, State University of New York, Upstate Medical University, Syracuse, New York; and ¶Department of Laboratory Medicine and Pathology, Mayo Clinic, Rochester, New York.

Disclosure: The authors declare no conflict of interest.

Address for correspondence: Barbara Robinson, Division of Cardiothoracic Surgery, State University of New York, Upstate Medical University, 750 East Adams Street, Syracuse, NY 13210. E-mail: anistasia36@yahoo.com.

DOI: 10.1097/JTO.0000000000000452

ISSN: 1556-0864/15/1004-0e18

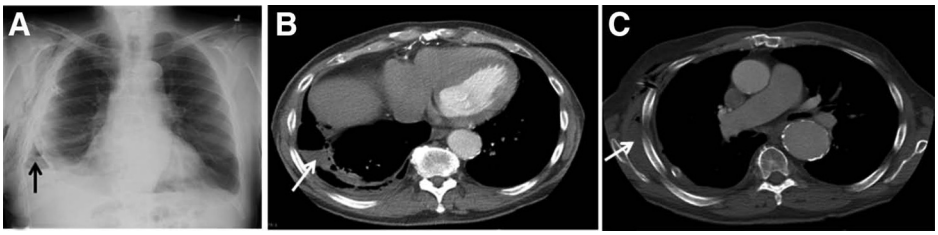

FIGURE 1. Imaging studies of the chest wall. *A*, Chest radiograph demonstrates a right-sided hydropneumothorax (arrow). *B*, CT of the thorax demonstrates a complex septated air-fluid collection (arrow) within the right pleural space. *C*, CT thorax showing a subtle elastofibroma tumor deep to the latissimus dorsi muscle (arrow). CT, computed tomography.

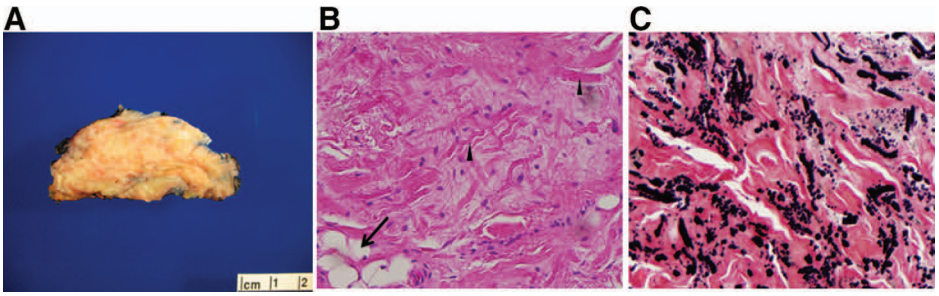

FIGURE 2. Immunohistochemical studies of the elastofibroma tumor. *A*, Gross image of right chest wall elastofibroma. *B*, H&E stain demonstrating dense, partly cellular collagenous tissue with entrapped adipose tissue (arrow) admixed with large, irregular, thick elastic fibers (arrowheads). The elastic fibers are deeply eosinophilic, branching, with areas of globules in a linear arrangement as "beads on a string." *C*, Elastin stain highlighting the fibers and the dense core and irregular margins of the globules, confirming the diagnosis of elastofibroma. H&E, hematoxylin and eosin.

Subsequent, intraoperative pleural peel cultures grew Gram-positive cocci in clusters; treatment with antibiotics for 6 weeks for chronic empyema resulted in resolution of symptoms, and he remains well 2 years postoperatively.

DISCUSSION

The etiology of elastoma remains unknown and may be a combination of similar acquired and genetic aberrations in the pathogenesis of elastofibromas and AS.[5] Elastofibromas are known to occur secondary to local mechanical friction; similarly, sheer stress had been implicated in the pathogenesis of AS.[4] The pathogenesis of elastofibroma also involves clonal fibrous proliferation, and CD34-positive endothelial progenitor cells were shown to be involved in the formation of some types of AS.[4] A clinical association between AS and *excessive* proteins as found in elastofibroma, which coexisted in our patient, has not been previously noted, whereas there is a known association between ascending aortic aneurysms and aortic valve insufficiency with *deficiency* of structural elastic proteins in some patients.[3] In summary, this is the first described clinical case of an association between elastofibroma and AS, with a potential similar acquired and/

TABLE 1. Patient, Age and Sex, and Location of Elastofibroma Dorsi in 158 Patients from 12 Series Published Since 2000

Year	References	Patients	Age, yr		Sex		Location of Tumor		
	First Author	*n*	Mean	Range	F, M	F:M	Right	Left	Bilat
2000	Schick[4]	3	58	53–63	1, 2	1:2	1	1	1
2001	Majo[5]	10	57	45–65	6, 4	1.5:1	5	1	4
2007	Daigeler–[6]	7	64	46–79	5, 2	2.5:1	4	2	1
2007	Gun[7]	7	54	44–85	7, 0	7:0	1	3	0
2007	Mortman[8]	6	—	—	3, 3	1:1	2	4	0
2008	Chandrasekar[9]	15	68	51–79	3, 12	1:4	7	6	2
2008	Muratori[10]	8	61	47–82	7, 1	7:1	4	3	1
2010	Koksel[11]	8	57	42–73	7, 1	7:1	2	1	5
2012	Ben Hassouna[12]	3	41	33–48	1, 2	1:2	0	3	0
2012	Cavallasca[13]	4	63	53–73	4, 0	4:0	2	1	1
2013	Lococo[14]	71	60	—	48, 23	2.1:1	34	25	12
2014	Karakurt[15]	16	61	38–78	11, 5	2.2:1	5	6	5
	Total	158	60	33–85	103, 55	1.9:1	67	56	32

or genetic etiology. Such a patient presenting with elastofibroma might prompt the clinician to be suspicious of multiple elastofibromas, and to listen for a systolic murmur in cases of AS and obtain an echocardiogram to search for valvular abnormalities, in this novel association between elastofibroma and AS. Although the etiology may be multifactorial, further research is necessary to investigate the common pathogenetic mechanisms.

REFERENCES

1. Jarvi O, Saxen E. Elastofibroma dorse. *Acta Pathol Microbiol Scand Suppl* 1961;51(Suppl 144):83–84.
2. Nielsen T, Sneppen O, Myhre-Jensen O, Daugaard S, Nørbaek J: Subscapular elastofibroma: a reactive pseudo tumor. *J Shoulder Elbow Surg* 1996;5:209–213.
3. David TE, Feindel CM, Bos J. Repair of the aortic valve in patients with aortic insufficiency and aortic root aneurysm. *J Thorac Cardiovasc Surg* 1995;109:345–351; discussion 351–352.
4. Gössl M, Khosla S, Zhang X, et al: Role of circulating osteogenic progenitor cells in calcific aortic stenosis. *J Am Coll Cardiol* 2012;60:1945–1953.
5. Ewart AK, Morris CA, Ensing GJ, et al. A human vascular disorder, supravalvular aortic stenosis, maps to chromosome 7. *Proc Natl Acad Sci U S A* 1993;90:3226–3230.
6. Schick S, Zembsch A, Gahleitner A, et al. Atypical appearance of elastofibroma dorsi on MRI: case reports and review of the literature. J Comput Assist Tomogr 2000;24:288–292.
7. Maj J, Gracia I, Doncel A, et al. Elastofibroma dorsi as a cause of shoulder pain or snapping scapula. Clin Orthop Relat Res 2001;388:200–204.
8. Daigeler A, Vogt PM, Busch K, et al. Elastofibroma dorsi–differential diagnosis in chest wall tumours. World J Surg Oncol. 2007;5:15–22.
9. Gun BD, Bahadir B, Behzatoglu K, et al.. Elastofibroma: a clinicopathologic and immunohistochemical study of seven cases and literature review. APMIS 2007;115:115–9.
10. Mortman KD, Hochheiser GM, Giblin, EM, et al. Elastofibroma dorsi: clinicopathologic review of 6 cases. Ann Thorac Surg 2007;83:1894–7.
11. Chandrasekar CR, Grimer R J, Carter SR, et al. Elastofibroma dorsi: an uncommon benign pseudotumour. Sarcoma 2008:2008:1–4.
12. Muratori F, Esposito M, Rosa F, et al. Elastofibroma dorsi: 8 case reports and a literature review. J Orthop Traumatol 2008;9:33–37.
13. Koksel O, Apaydin FD, Ayan E, et al. Elastofibroma dorsi: review of eight cases. Surg Today 2010;40:423–7.
14. Ben Hassouna J, Hamdi N, Ben Bachouche B, et al. Elastofibroma dorsi. Orthop Traumatol Surg Res 2010;96:717–720.
15. Cavallasca JA, Sohn DI, Borgia AR, et al. Elastofibroma dorsi: review of 4 cases. Reumatol Clin 2012;8:358–360.
16. Lococo F, Cesario A, Mattei F, et al. Elastofibroma dorsi: clinicopathological analysis of 71 cases. Thorac Cardiovasc Surg. 2013;61:215–22.
17. Karakurt O, Kaplan T, Gunal N, et al. Interact Cardiovasc Thorac Surg. 2014;18:197–201.